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ON THE ETIOLOGY OF "SCOTTY CRAMP" AND "SPLAY" — TWO MOTORING DISORDERS COMMON IN THE SCOTTISH TERRIER BREED

By

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ANDERSSON, B. and M. ANDERSSON: *On the etiology of „Scotty Cramp" and "Splay" — two motoric disorders common in the Scottish Terrier breed.* Acta vet. scand. 1982, 23, 550—558. — The skull-cap, the meninges, and the dorsal surface of the cerebral cortex were inspected in 20 Scottish Terriers subjected to euthanasia because of severe Scotty Cramp, and in 1 Cocker Spaniel and 2 Scottish Terrier puppies sacrificed because of Splay (inability to stand on the hind legs).

Apparent abnormalities observed in the 12 younger (6 weeks to 18 months old) Scotty Cramp animals were firm medial adhesion of the dura to the skull-cap (all animals), meningeal hemorrhage (10 animals), and bony jags on the inner side of the skull-cap making impressions in the underlying dura and cerebral cortex (8 animals).

Similar bony irregularities causing dural and cortical impressions were observed in 5 out of 8 Scotty Cramp dogs belonging to a senior age group (3 to 11 years old). Meningeal hemorrhage was observed only in 1 animal belonging to this group, but consistent observations were scarified appearance of the dura along the midline and induration around superior cerebral veins medially in the cruciate sulcus.

Meningeal hemorrhage and firm medial adhesion of the dura to the skull-cap were consistent observations in the Splay puppies (6 weeks to 3 months old).

The possibility is discussed that the ultimate cause of Scotty Cramp and Splay may be abnormalities in the development of the parietal and frontal bones, of their interconnection, and of their relation to the underlying meninges. This in turn may exert mechanical, circulatory or other influences causing dysfunction of the cerebral motor cortex.

Scotty Cramp; Splay; Scottish Terrier; meningeal hemorrhage; skull-bone abnormalities.

The episodic motoric disorder commonly known as Scotty Cramp was originally studied in Scottish Terriers 40 years ago by *Klarenbeek et al.* (1942). That, and subsequent investigations on Scotty Cramp (*Smythe* 1945, *Meyers et al.* 1969, 1971) provide a rather uniform picture of the development and the symptoms of the disease. The motoric disturbance generally makes its début at ages between 2 and 6 months, and the symptoms then usually become apparent when the dog is physically strained or excited. The symptoms consist of transient muscular hypertonicity preferably involving the muscles of the hind legs and the pelvic region. As a result the back becomes arched and the dog gallops like a hare. The duration of the fits may vary from less than 1 min to ½ h. *Klarenbeek et al.* (1942) pointed out that disposition for the disease by all probability is hereditary. This has later been conclusively shown by *Meyers et al.* (1970). All investigators of Scotty Cramp agree upon that the intermittent cramps are due to some central nervous system (CNS) dysfunction. However, with one exception (*Andersson & Åkerlund* 1976), previous morphological examinations have not disclosed any CNS abnormalities which could explain the transient muscular hypertonicity characterizing the disease.

Another motoric disorder, rather common among Scottish Terrier puppies, is so-called Splay or Splay-leg. As regards dogs, Splay has to our knowledge not been dealt with in any veterinary medical publication, but has now and then been discussed in breeders' periodicals (i.e. West of England Scottish Terrier Club Bulletin, Nos. 159 and 160, 1975). The severeness and the duration of this motoric disturbance may vary considerably. Normally a Scottish Terrier puppy (like puppies of other breeds) is capable of standing on all 4 legs at the age of about 2 weeks, and runs around 1 or 2 weeks later. At the corresponding ages puppies affected with Splay are unable to control the movements of their hind legs and remain resting on the belly using only the front legs for locomotion. Independently of any kind of external support imposed upon them, most Splay puppies get up on all 4 legs before the age of 8 weeks, and soon afterwards develop normal faculty of locomotion. However, some puppies with more severe Splay remain unable to get up on the hind legs; this sooner or later leads to patellar dislocation, depression of the sternum and deformation of the pelvis.

Thanks to the far-sighted and positive attitude of Swedish dog-owners and -breeders we have over the past several years had the opportunity to examine brains, meninges and skull-caps from Scotty Cramp and Splay dogs subjected to euthanasia. Some of the observations made were briefly reported previously (*Andersson & Åkerlund* 1976). Here is presented the integrated result of the examinations, which suggests that both Scotty Cramp and Splay may be manifestations of motor cortex dysfunction elicited by abnormal development of the skull bones.

MATERIAL AND METHODS

Scotty Cramp dogs

Necropsy material was obtained from 20 Scottish Terriers which had been subjected to euthanasia (intramuscular azopromazine followed by intravenous pentobarbitone) because of severe Scotty Cramp. Twelve of the dogs (8 ♀, 4 ♂) were subjected to euthanasia at ages between 1.5 and 18 months. In 1 of the animals (age 5 months) belonging to this younger group the head was perfused via the carotid arteries with isotonic saline followed by 6 % formaldehyde solution immediately after death. The older group (ages 3 to 11 years) consisted of 8 dogs (4 ♀, 4 ♂). Pre-necropsy perfusion of the head was performed in 2 animals belonging to that age group.

Splay dogs

Necropsy material was obtained from 1 male and 1 female Scottish Terrier puppy (ages 7 weeks and 3 months, respectively) and from 1 Cocker Spaniel puppy (♀, age 7 weeks) affected with severe Splay. Pre-necropsy perfusion of the head was not performed in any of the Splay puppies.

Necropsy procedure

Post-mortem examination was performed as soon as possible (in the majority of cases within 1 h) after euthanasia. After removal of covering skin and muscular tissue a dental drill was used to make an oval bilateral section through the frontal and parietal bones with care taken to avoid damage to the underlying dura. The skull-cap was then gently lifted and separated from the adherent dura. The inner surface of the skull-cap and the dura (in situ) was inspected before the surface of the cerebral

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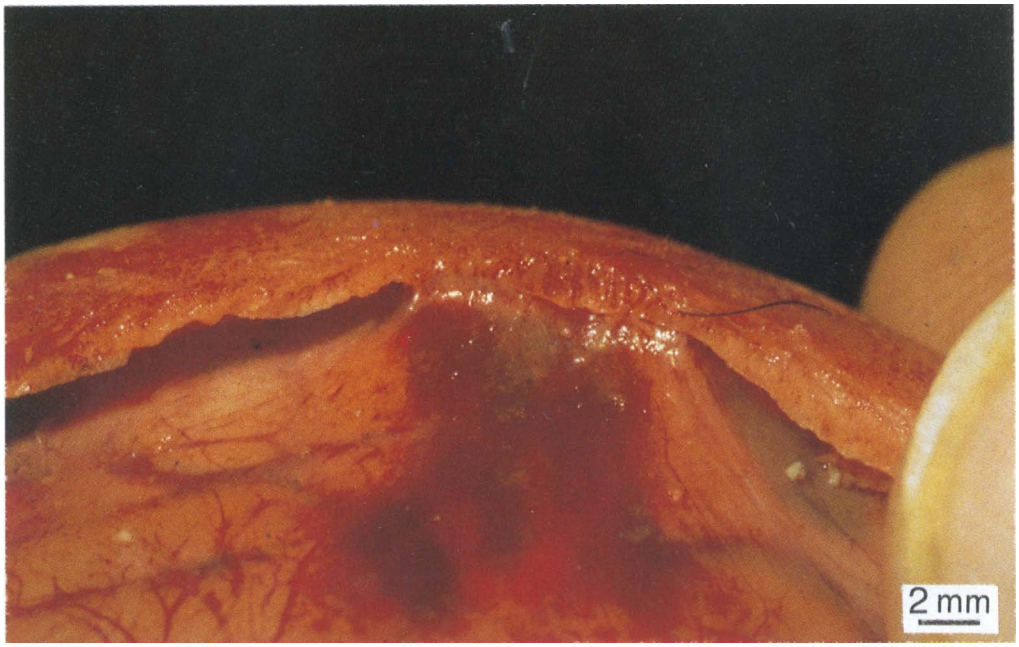


Figure 1.
Firm midline adhesion of the dura to the skull-cap and meningeal hemorrhage observed in the youngest (6 week old) Scottish Terrier subjected to euthanasia because of Scotty Cramp. Two fingers lifting the skull-cap are seen to the right.

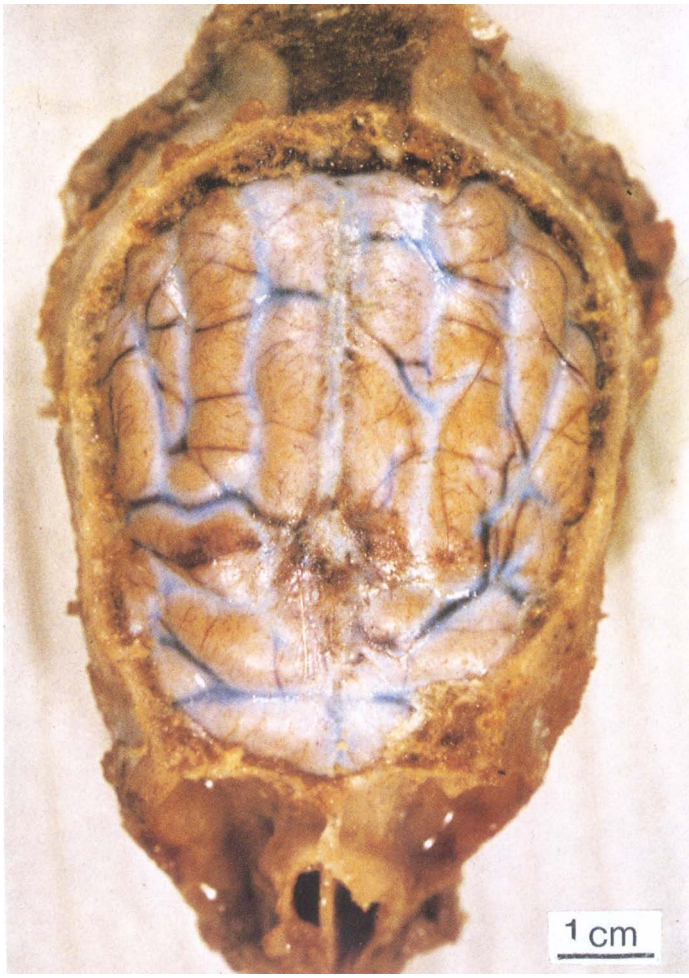


Figure 2.
The exposed dura (after formaldehyde fixation) in a 10 months old Scotty Cramp dog showing chronic scarified appearance of the dura underneath the crossing of the sagittal and parie-frontal bone sutures.

Figure 3.
Meningeal hemorrhage in the 3 months old Scottish Terrier puppy affected with Splay. The hemorrhage is extending backwards along the midline from the level of the cruciate sulci and is also seen bilaterally (underneath the parie-frontal bone suture).

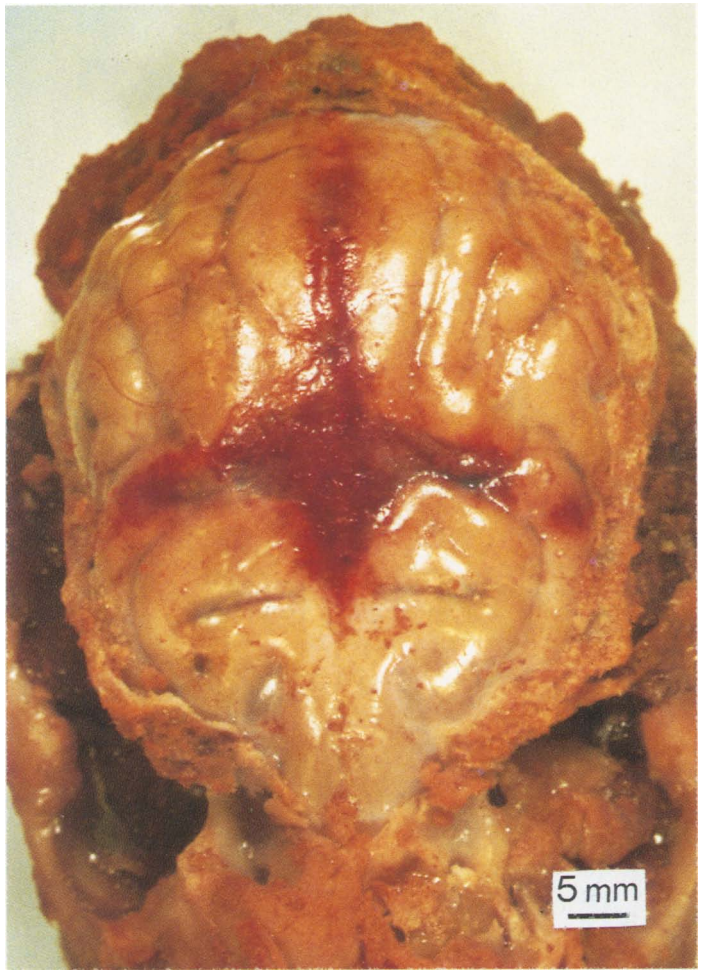
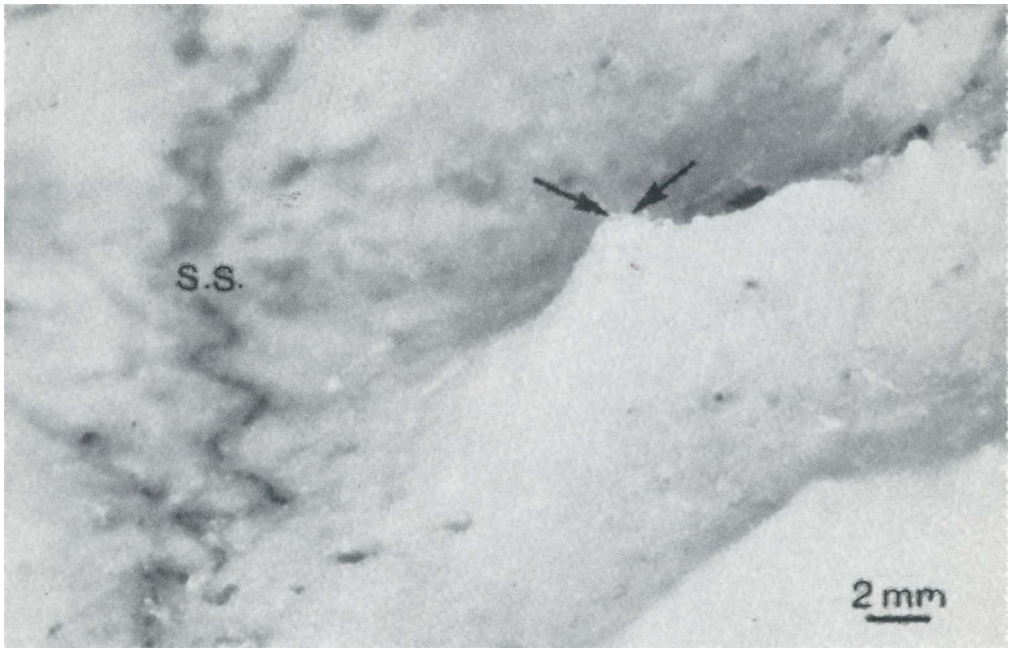


Figure 4.
The inner surface of the frontal bone in a 7 months old Scotty Cramp dog showing (on the right side of the midline at arrows) a bony jag which had made an impression in the dura and underlying motor cortex (the post-cruciate gyrus). Cramping had made its debut 1 week before the animal was subjected to euthanasia and no meningeal hemorrhage was observed in the dog.
S.S. = Sagittal bone suture.



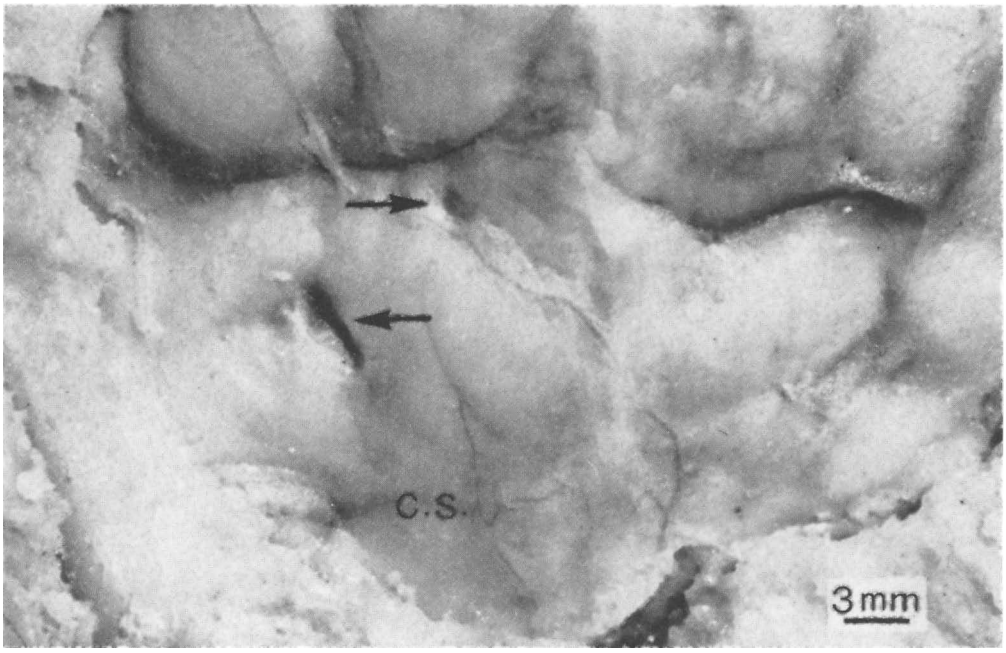


Figure 5.
Impressions in the dura and the cerebral cortex (within and at the posterior border of the right postcruciate gyrus) caused by jags on the inside of the frontal bone of a 7 years old Scotty Cramp dog. The impressions are marked by arrows.
C.S. = Transverse level of the cruciate sulcus.

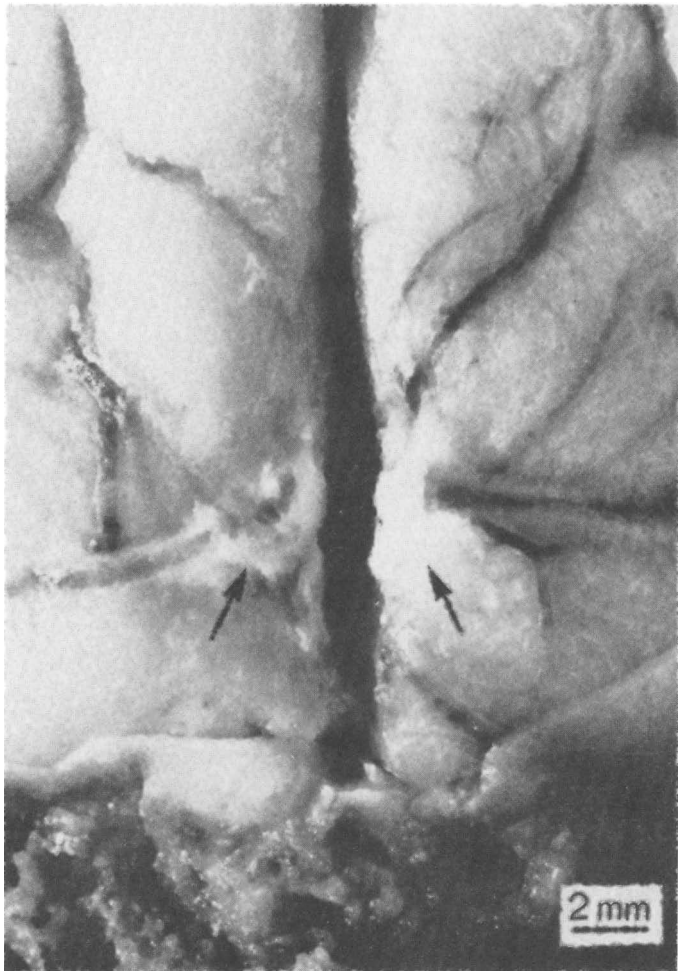


Figure 6.
Bilateral indurations (at arrows) around superior cerebral veins medially in the cruciate sulci of a 7 years old Scottish Terrier affected with Scotty Cramp.

cortex was exposed by removing the dura-arachnoidea. Removal of these meninges was in most cases not made until the material had been preserved in 6 % formaldehyde solution for a week or longer.

RESULTS

Observations in Scotty Cramp dogs

Age group 6 weeks to 18 months. In all animals belonging to this age group the dura was firmly adherent to the skull-cap along the sagittal bone suture (Fig. 1). In 6 of the dogs transverse adhesions were also observed along the parie-frontal bone suture. In all but 2 dogs meningeal bleedings or blood congestion were observed medially behind the level of the cruciate sulcus and/or bilaterally underneath the parie-frontal bone suture (Figs. 1 and 2). Exceptions were the dog in which the head had been perfused before necropsy, and a 7 months old animal where the cramping had made its début as late as 1 week before euthanasia. In the former animal the dura had a scarified appearance under the crossing of the sagittal and parie-frontal bone sutures, and behind this level the sagittal sinus remained over-filled with blood in spite of the fact that the head had been perfused. In the latter animal (late cramp début) a drip-stone-like jag (Fig. 4) on the inner surface of the frontal bone had made an impression in the dura and cerebral cortex about 5 mm behind the right cruciate sulcus. Similar bony jags or puckers making more or less pronounced impressions in the dura and the underlying cerebral cortex were observed also in 7 other dogs belonging to the junior age group.

Age group 3 to 11 years. Also in the senior age group the dura was firmly adherent to the skull-cap along the midline. A circular meningeal hemorrhage was observed underneath the crossing of the sagittal and parie-frontal bone sutures in 1 of the dogs (age 5 years). Meningeal bleedings were not seen in the other 7 animals, but here the dura appeared thickened and scarified at sites where bleedings and blood congestion were observed in the Scotty Cramp dogs belonging to the junior age group. Drip-stone-like jags and puckers on the inside of the skull-cap in front of and behind the parie-frontal bone suture were consistent findings in the senior age group. These bony irregularities were most pronounced in the 4 oldest animals (7 to

11 years) and had in 5 dogs made impressions in the dura and the underlying cerebral cortex near behind the cruciate sulcus (Fig. 5). Another consistent observation in the senior age group was uni- or bilateral induration medially in the cruciate sulci around superior cerebral veins emerging into the sagittal sinus (Fig. 6).

Observations in Splay puppies

Firm adhesion of the dura to the skull-cap and pronounced meningeal hemorrhages were observed in the 3 Splay puppies at the same sites as in the Scotty Cramp dogs belonging to the junior age group. An additional observation in the Cocker Spaniel puppy was conspicuous blood congestion in the sagittal sinus behind the level of the sagittal and parie-frontal bone suture crossing. Fig. 3 shows the extensive meningeal hemorrhage which had occurred in the 3 months old Scottish Terrier affected with Splay. Here the hemorrhage also extender bilaterally underneath the parie-frontal bone suture.

DISCUSSION

The thorough investigations of Scotty Cramp performed by *Meyers et al.* (1969, 1971) have eliminated abnormalities in general metabolism, skeletal muscular function, and neuromuscular transmission as possible etiological factors, and have lead to the conclusion that the crucial defect resides in the CNS motoric system. Since at the same time these investigations have failed to reveal morphological CNS abnormalities, *Meyers et al.* (1973) have suggested and produced indirect evidence that impaired production of the neurotransmitter serotonin (5-hydroxytryptamine, 5-HT) might be the ultimate cause of Scotty Cramp. In support of the hypothesis are observations that treatment of Scotty Cramps dogs with agents increasing the cerebral serotonin concentration have a beneficial effect whereas agents decreasing the CNS concentration of the neurotransmitter aggravate the clinical signs (*Peters & Meyers* 1977). What seems to obscure the serotonin hypothesis, however, is that dogs affected with Scotty Cramp have been found to have normal brain content of serotonin (*Meyers & Schaub* 1974). Furthermore, it appears hard to conceive that impaired cerebral production of serotonin should become manifested solely as intermittent skeletal muscu-

lar hypertonicity generally involving only the posterior part of the animal. The role played by CNS serotoninergic neurons in the control of motor activity is obviously complex and cerebral serotoninergic pathways are apparently also involved in emotional and homeostatic mechanisms (*Gerson & Baldessarini* 1980). Therefore, impaired CNS serotonin production would be expected to become manifested in a much more complex manner than simply as episodic spasms affecting a restricted group of skeletal muscles. It appears more likely that the abnormality leading to Scotty Cramp mainly affects the area of the motor cortex which is responsible for voluntary control of the muscle groups displaying hypertonicity, or affects subcortical projections from that area.

The voluntary control of limb and trunk movements in the dog is primarily exerted from the postcruciate gyrus in which the posterior parts of the body are represented most medially (*Breazile & Thompson* 1967). This is also the part of the motor cortex which in the first hand might have displayed dysfunction in consequence of the apparent abnormalities observed in the present dogs. In 10 out of 12 Scotty Cramp dogs belonging to the junior age group, and in the 3 Splay puppies, medially located meningeal hemorrhage was present at, and behind the postcruciate gyrus, and in some animals the hemorrhage also extended laterally underneath the parie-frontal bone suture (Fig. 3). Constant findings in the Scotty Cramp dogs belonging to the senior age group were scarified appearance of the dura along the midline (suggesting earlier meningeal hemorrhage), and induration around superior cerebral veins medially in the cruciate sulcus (Fig. 6). In a majority of animals belonging to both age groups bone jags or puckers had made impressions in the dura and the underlying motor and/or somatosensory cortex. It seems conceivable that the meningeal hemorrhage and scarification as well as the bony irregularities could have mechanically affected the motor cortex and in that manner could have acted epileptogenic in areas controlling the movements of the hind quarter. Meningeal hemorrhage and the induration seen medially in the cruciate sulci may also have impaired the drainage of blood from motor and somatosensory parts of the cerebral cortex. Such mechanical and circulatory effects may have become accentuated when the brain volume increased in response to elevated arterial blood pressure during physical strain and emotion. This

could provide an explanation for the fact that Scotty Cramp usually becomes manifested during exercise and excitement. In the Splay puppies it seems conceivable that the pronounced meningeal hemorrhage (Fig. 3) by mechanically affecting medial parts of the motor cortex or by impairing its blood circulation may have prevented normal voluntary control of the hind limb movements.

It has for obvious reasons not been possible to obtain necropsy control material from young healthy Scottish Terriers. To our knowledge, dural adhesion to the skull-cap and puckers or jags on the inside of the parietal and frontal bones are not uncommon post-mortem findings in older dogs of various breeds. However, the fact that such bony irregularities and meningeal hemorrhage were present in 6 weeks to 18 months old Scotty Cramp dogs suggests inherited abnormalities in the development of the skull bones, of their interconnection, and of their relation to the underlying dura. The reason why particularly dogs belonging to the Scottish Terrier breed should be predisposed for these abnormalities appears puzzling. However, typical Scotty Cramp has been reported also in Cairn Terriers (*De Lahunta* 1977), the present material includes a Splay puppy of the Cocker Spaniel breed, and Splay is not uncommon among Swedish Vallhund puppies (breeders' communication). It shows that the 2 motoric disorders discussed here are not exclusively confined to Scottish Terriers. Of comparative neurological interest are — as pointed out by *Meyers et al.* (1971) — the striking clinical similarities between Scotty Cramp and the Stiff-man syndrome which is a very rare motoric disorder in humans (*Gordon et al.* 1967). Like Scotty Cramp, the Stiff-man syndrome consists of episodic muscular hypertonicity often restricted to posterior skeletal muscle groups and triggered by exercise and excitation. It has so far not been possible to distinguish any particular CNS defect causing the Stiff-man syndrome. However, of some interest with regard to the present observations in Scotty Cramp dogs appears to be the recent tomographic demonstration of frontal interhemispheric fissure dilatation in a case of Stiff-man syndrome (*Maida et al.* 1980).

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SAMMANFATTNING

Om orsaken till „Skottekramp" och „Splay" — två hos skotsk terrier vanliga motoriska störningar.

Skalltak, hjärnhinnor och dorsala hjärnbarken har undersökts makroskopiskt hos 20 skotska terrier som måst avlivas för svår skottekramp, samt hos en cocker spaniel valp och 2 skotska terrier valpar vilka måst avlivas för splay (oförmåga att resa sig på bakbenen).

Synbara abnormiteter hos 12 yngre (6 veckor till 18 månader gamla) skottekrampshundar var uttalad medial sammanväxning av duran med skalltaket (samtliga djur), hjärnhinneblödningar (10 djur) och bentaggar på insidan av skalltaket, vilka orsakat intryckningar i dura och hjärnbark (8 djur).

Motsvarande bentaggsintryckningar sågs hos 5 av 8 äldre (3 till 11 år gamla) skottekrampshundar. Medial hjärnhinneblödning observerades endast hos en hund i den äldre gruppen, men ärrvandling i duran under den sagittala bensuturen hos övriga djur tydde på att dessa tidigare varit utsatta för hjärnhinneblödningar. Vidare sågs hos samtliga äldre djur indurativa förändringar runt de ytliga cerebralvenerna medialt i Sulcus cruciatus.

Uttalad medial sammanväxning av duran med skalltaket och hjärnhinneblödningar observerades hos de 3 splay-valparna (ålder 7 veckor till 3 månader).

Möjligheten diskuteras att den primära orsaken till såväl skottekramp som splay må vara en onormal utveckling av frontal- och parietalbenen, av deras sammanfogning och relation till underliggande hjärnhinnor, och att detta i sin tur leder till mekanisk, cirkulatorisk eller annan påverkan på den motoriska hjärnbarken med rörelsestörningar som följd.

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